

**Barbara Barletta**

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**From:** [REDACTED]  
**Sent:** Wednesday, May 04, 2016 11:25 AM  
**To:** Barbara Barletta  
**Subject:** RE: Consent decree - One parent's nightmare  
**Attachments:** BHDDH Complaint (Revisions).docx; Physician Advisory Letter Jan 2008.pdf; 160329131811\_0001.pdf; Our Donated automobile.jpg

Dear Honorable Judge J. McConnell, Jr.

It's been a particularly hard two days for my family.

Yesterday, my daughter, Christine [REDACTED] (who is a young adult with significant special needs) was taken to the Newport Hospital ER room. She had insomnia the night before and was up most of the night. I am of the mindset that her group home agency's administration had thought she would be admitted, but the treating doctor saw no medical or clinical reason to admit here. However, the group staff has left the hospital under orders from the their supervisors. Shortly thereafter, my wife received a call from the Director of Residential Services, stating that Christine **could not return to the group home**. She was left at the hospital. Apparently, staff members were told to have no contact with the family.

Your Honor, I truly believe this was a retaliatory measure in response to a grievance that I had filed on April 27, 2016 against the group home and day program provider. Attached, you will find the original letter that was presented to BHDDH on April 27, 2016. It will provide help fill in much of necessary background. I do apologize for its length. You will also find some attached applicable files that were attached to my original letter.

I understand that Your Honor serves a particular purpose and that is to weight the facts concerning the case of the U.S. Department of Justice and The State of Rhode Island as it applies to the Consent Decree. I guess the main reason I write you is simply to point out the type of obstacles parents and guardians of our loved ones with developmental disabilities face each and every day. I do not know what the future holds for Christine and my family, but I feel strongly that the "system" (rather intentionally or unintentionally) has wronged one of our most vulnerable members of our society

Your Honor, thank you for your time. And thoughtful consideration

Sincerely,

Steven [REDACTED]

[REDACTED]  
Cranston, RI 02921-2714  
April 27, 2016

Office of the Director  
Department of Behavioral Healthcare,  
Developmental Disabilities and Hospitals  
Barry Hall  
14 Harrington Road  
Cranston, RI 02920

Director Montanaro:

My wife, Jo-Ann, and I are the biological parents to three boys and the adoptive parents of three girls. All of our girls have some level of special need. In the late 1980's, we became foster parents, which led to the adoption of our girls. Our three biological boys (an attorney, a nurse and a pharmacist) are grown and living on their own. It seems that our girls will always need our assistance.

When our girls were much younger, we found much of our time was spent navigating the world of special education. My wife and I had lived through, helped reform, and witnessed major changes in the way academic services were delivered to students with special needs. In those years, I thought the world of special education was the absolute worse. I was wrong. Now we are fully immersed in the world of advocating for our adults daughter with special needs and the tragedy that is our state's version of helping and servicing adults with special needs. (I willingly admit that I thought long and hard about typing that last sentence, as I truly do fear repercussions.)

Our middle daughter, Christine was born prematurely on [REDACTED]. My understanding is that, at one point, she had stopped breathing, but she was resuscitated. Many years later, testing showed that she was a young lady with a Chromosome 15q Duplication Syndrome. The manifestations of this disorder are characterized by autistic like behavior, Obsessive-Compulsive disorder and Tourette's. She is also at risk for seizures and sudden death. (See attached document)

In December of 2012, Christine was hospitalized at Butler Hospital. Christine had turned 21 in July of 2012 and she was extremely unhappy with her life; there was a tremendous drop in services when she transitioned from adolescence to adulthood. She self-dialed 911 and she was taken to RI Hospital and later transferred to Butler Hospital. While at Butler, she was administered the maximum dosage of the medications Tegretol and Anafranil in a relatively short period of time. This caused her liver to go into failure and our family spent New Year Day 2013 at her bedside at Kent County Hospital wondering if she would need a liver transplant.

Based on these events, we reached out to the Department of Behavioral Healthcare, Developmental Disabilities and Hospitals to seek their guidance. Christine's doctors, BHDDH and administrators from other agencies that had worked with our family at that time all recommended that Christine should be placed in a group home. Going along with this proposal was certainly a bittersweet decision made by my wife and I. While we love our daughter unconditionally, being her caregiver 24/7 was extremely draining. Her caregivers have to be considerably patient and "on their game" always. Christine very rarely sleeps entirely through the night and the amount of times she gets up will vary depending on her state of anxiousness. We hoped that having fresh staff available every eight hours would enhance her quality-of-life. At the very least, her caregiver would be less apt to burn out. There is no doubt that Christine is a lot of work. My wife and I realize how challenging she truly is as we have been her only caregivers for 21 years.

I believe the long process of identifying a group home for Christine began in earnest in the early months of 2013. To our surprise, in March of 2013, a group home was identified. It was an all-women group home located in Newport, RI operated by the James L. Maher Center and the home had a single opening at that time. We visited the facility without Christine being present. We wanted to be sure that this would be a suitable home for our daughter. Christine's state appointed social worker visited the home as well as Christine's day program one-on-one aide who would help with the eventual transition. Everyone involved with Christine's case wanted the transition to be appropriate, successful and smooth. This was a seemingly well thought out scenario.

Once we were sure that the state was on board (through conversations with Christine's social worker), and that the group home was on board (they said they wanted Christine to be part of their clientele), the application process began. We were led to believe that Christine would be able to move into her new home by July or August 2013. We only needed the approval of the application by the "higher ups" at the Department of Behavioral Healthcare, Developmental Disabilities.

We eventually received verbal approval from Christine's social worker, as well as the Maher Center group home coordinator, that the application had been approved. We were only waiting for a move-in date. We were told that there was a young lady who had been placed in the home on a "temporary 45 day placement" currently occupied Christine's spot. As soon as the young lady was placed elsewhere, Christine could move in. This was presented as an after-thought ... not at all as a scenario that would nix Christine's placement.

We took Christine to visit the Newport group home after we received the verbal approval of her application. These are the necessary baby steps we take whenever we introduce Christine to something new.

Skip to January 2014: we were still being told, "Christine will be placed at the Newport group home. However, the individual with the '45 day temporary placement' has still not been placed elsewhere, so Christine is not able to move in yet."

On January 21, 2014, at the urging of several individuals, I contacted a "higher-up" at the Department of Behavioral Healthcare. He said he was familiar with the case and stated that we should not have been told that the placement of the young lady currently living in the Newport group home was "temporary." He offered no resolution other than that we could start the process of identifying another possible group home for Christine if we wanted to. This was being presented like this was a new option but Christine's name had already been re-entered on the list because of the amount of time that had already passed. No other agency had expressed an interest. I expressed to him that a lot of time and effort was put into identifying the group home in Newport and that we did not even discuss it with Christine or allow her to visit it until we were led to believe that the placement would happen.

Finally, on August 1, 2015, more than a year and a half after we had began looking, Christine moved into the group home called the Jimari House in Newport. When she was living at our home, and with supports in place, Christine had become a volunteer as a "play partner" at the Hasbro's Big Back Yard in Roger Williams Park. We wanted Christine to continue to volunteer at the zoo and we were told by a representative of the Maher Center that, because she qualified for one-on-one support, "there would be no reason why she couldn't go out anytime she wanted."

In addition to having Christine live at Maher owned group home, we decided to also utilize the James L. Maher Center for her day placement, believing that there would be a healthy collaboration between the staff of Christine's group home and the staff of the day program.

When the day came to sign the papers that allowed Christine to live in her Newport Group home and attend The James. L. Maher Center day car program, we were presented with a caveat to the plan of continuing to allow her to volunteer at the zoo: transportation would be an issue; there were only so many vehicles available. My wife and I so desperately wanted this new placement to work that we would do practically anything in our power to ensure its success. To solve the possible transportation issue, we donated a car to the Maher Center. This letter, dated July 24, 2014, was presented along with the paperwork transferring title of our automobile to the agency:

To Whom It May Concern:

I, the undersigned, do hereby swear/affirm the following:

That I, Steven E. [REDACTED] Cranston, RI 02921-2714, am **GIFTING** a 2004 Toyota Corolla LE, Vehicle Identification Number [REDACTED] **The; James L. Maher Center; 120 Hillside Avenue; Newport, Rhode Island 02840** for no monetary consideration.

The reason for this gift transaction is because my daughter, with Special Needs, is a resident of this group home and it is important for me that the staff of her group home has immediate access to a dependable vehicle for her transportation.

This declaration is made with my full knowledge and understanding and under no duress.

Sincerely,  
Steven [REDACTED]

The best laid plans ....

Sadly, her placement has not been what I would deem "successful." She no longer volunteers at Roger Williams Zoo and hasn't been allowed to for many months. Her day program will no longer transport her there. The reasons we are given range from lack of adequately trained staff to the client's unsafe behavior.

Christine has been taken to the Newport emergency room on many occasions due to her behaviors. Sadly, she has been at an emergency room more in the year and a half she was in her group home than she has been in all the time she lived with us prior. We are told, "It's protocol."

At one meeting (where Christine's social worker from the state and staff from the Maher Center were present), I voiced my belief that if we did not agree to medicate/sedate her, I had the absolute feeling that the James L. Maher Center would give notice on Christine. Indeed, on one of those occasions where Christine was taken to the Newport Emergency Room (and subsequently hospitalized there), we indeed received a thirty-day termination of services notice from the executive director of the James L. Maher Center. Jo-Ann and I are not anti-medication. However, it is well documented that medications pose a grave risk for Christine's liver functions. Further, there are studies that show that medications that usually have some success with those who have autism don't have the desired effect on individuals with those who have Chromosome 15q Duplication Syndrome. We have always found that the best medication for Christine is behavior modification. We have found that it is important to redirect Christine's behavior before it escalates to the point of her needing to be restrained. We are not anti-restraint, but we have found that restraining Christine heightens her anxiety and she tends to fight back harder, rather than calming down.

Present-day Christine is medicated ... daily ... with PRN's.\* Lots and lots of PRN's. At the request of the Maher administration, Christine's doctor recently prescribed a different PRN that could be dispensed after she had already swallowed two doses of her regular PRN. Christine now has to have her liver function levels tested regularly due to the amount of medications she is taking. This requires taking her to a lab to have her blood drawn. Worse, rather than achieving the desired effect of controlling her behaviors, her daily dose of medications makes her very tired during the day and, in turn, she becomes more anxious and more agitated than she ever had before.

On March 24, 2016, we forwarded the following email to the nurse who is employed by the James L. Maher Center:

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\* The initials, PRN, stand for "pro re nata," a Latin phrase that roughly translates to "as needed" or "as the situation arises."

I am growing concerned about the increasing number of PRN's Christine is being given daily across both the day program and in the home. Christine is no longer able to talk to me on the phone the way she had in the past. She is constantly yawning and obsessing about events that she used to do with her family when she lived at home. She used to get excited when she spoke about what she did at the day program, but now when I asked her about her day, she repeats, "I don't like PRN's." The last time Christine was excited about the day program was when Josette told her she had an option to work at Save the Bay, which has since been nixed.

While I realize that PRN's may be necessary at times, I'm not sure I fully understand the protocol of when staff should be administering a PRN. What signals a staff person that there is a need for a PRN? Which drug is given first and what the guidelines are for administration of them? I would like a clarification on this please.

As her legal guardian, I am requesting that I be provided with a record of dates and times that PRN's have been administered to Christine. I would also like to know the TOTAL number of PRN's (in pills and times) that have been administered to Christine since they were first prescribed. In addition to the total, I would like them separated into two groups: those administered during the day program and those that are given in the group home. I would like to know how often they are given to her across both the workshop and her home. I believe Dr. Klein began approving them in December. I would like this list no later than March 31, 2016.

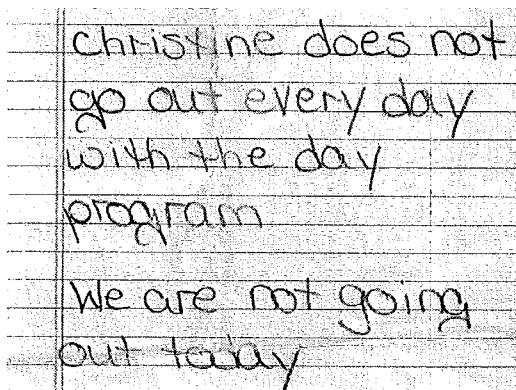
Of another importance is trying to get to the bottom of Christine's escalating behavior that warrants the need for more and more PRN's. Unfortunately that can be so difficult because she isn't able to express her feelings in a typical manner, but rather through her behavior. If her behavior is increasing in agitation I believe we need to look into what may be attributing to it. As always I remain concerned on how her liver will process the combination of medications and how medication is affecting her ability to participate in activities of daily life.

I look forward to working together with you in regards to Christine's health concerns.

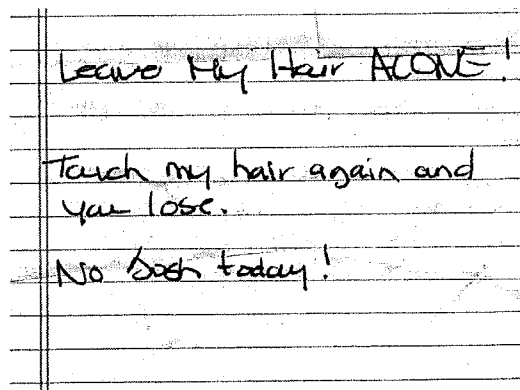
Thank you,  
Jo-Ann [REDACTED]

As of this date, we have not received this list from the Maher Center. On March 31, 2016, we were told that it was being "working on it," but as of the date of this letter, we have not received anything. Neither have we heard anything. The daily dispensing of her medication continues and remains uncurbed.

On another occasion, we had the opportunity to go through Christine's backpack, an item that our daughter carries with her everywhere. We found several notes inside the backpack that raised our concern, including the ones copied below:



christine does not  
go out every day  
with the day  
program  
  
We are not going  
out today



Leave My Hair Alone!  
  
Touch my hair again and  
you lose.  
  
No Sash today!

Members of Christine's staff wrote these notes. I was drawn to the use of the terms "does not go out every day with the day program" and "lose." Other notes show that Christine is threatened with not being allowed to attend events that are two days or more away if she does not control her behavior. Controlling inappropriate behavior can be a difficult task for adults with autism. I think it is fair to say that adults with autism live in a 15-minute world. To lose planned outings that are days away is punitive. We sent copies of these notes to the administration. We received no response, but it is interesting to note that Christine's backpack is always cleaned out now.

Based on documents we recently signed,\* it seems that The Maher Center may bill for one-on-one staffing for 60% of our daughter's day but the reality is that Christine rarely leaves the building. We are told that the manager of the day program insists that two-on-one staff accompany Christine when she goes out in the community and that combination is rarely available. Christine is regularly omitted from planned trips due to her level of care and "not enough room in the van." Most days, she is made to watch a movie in her day program, an activity that was not one of her favorite things to engage in.

When Christine was living at our home, we spent a lot of time taking her out into the community. She learned how to order food from a menu at restaurants. We took her to movies and we taught her how to bowl. She is a RI Special Olympian who participates annually at Rhode Island's summer games at the University of Rhode Island. She loves to sing and she plays the piano by ear. She wants to be a chef in "real life" and often asks about attending Johnson and Wales renowned culinary school. She loves sciences, the solar system and the universe. She wants to help care for young children, especially those confined to a wheel chair. She is intrigued with most things historical, including historical sites and artifacts.

There is a beach that is literally a five-minute walk from Christine's group home. Christine knows how to swim, as this is another skill that has been taught to her. Yet, neither her day program staff, nor her group home staff, takes her there. We purchased her a membership to Save-The-Bay, which is located driving minutes from her group home. This is one of Christine's favorite places to visit and it had even been mentioned as a possible site for her to do volunteer work, but her day program manager vetoed the idea of having her volunteer there. Like the beach, Jo-Ann and I have taken her there more than her staff has.

Christine doesn't like the noise that fluorescent lights generate and she doesn't like the sound of an air conditioner's compressor. She does not like to touch the fur of animals or stuff toys, but she loves reptiles, fish and snakes. Christine has obsessive-compulsive disorder and will ask the same question over and over again. In fact, her normal pattern of communication is asking questions and not making statements. She has language, but she does not know how to use words to express when she is anxious or unhappy. She will not tell you when she is hungry, but she loves all types of foods and she does get hungry.

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\* See attached document that reads "Quarterly Purchase Order for Individuals Receiving 24 Hour Residential Supports."

Christine has Tourette's Syndrome and she will make noises that she cannot control. She likes to sleep wrapped in blankets, as if she is mummified.

Christine grew up in a home where she had access to computers. She is very Internet savvy and enjoys using Disney applications on her Apple iPad. For the most part, however, she is not allowed to engage in this activity at her group home because they will not allow her access to the Internet. I say "for the most part," because, once in a while, certain staff members will take pity and allow her limited access. Once again, she has an ability that she is not allowed to use. Shouldn't her world be about choices?

We were told by the administration of the James L. Maher Center that Christine would be the first of a new younger, population of adults with autism at the Jimari Group home location on Carroll Avenue in Newport, RI. Within six months of her placement, we were informed that The Maher center had abandoned those plans and they were forced to sell another of their group homes due to monetary issues.

I believe this is one of the many reasons for my daughter's unsuccessful placement. From my observation, the majority of the residents of her group home, and those of the Maher Center day program, are much older and more sedentary than my daughter. Once the purported plans for the introduction of a younger adult population with autism was abandoned, the James L. Maher Center has seemingly not bought into the education and training needed to enhance the lives of their current population who live with autism. For example, my wife has provided and introduced many materials (i.e.: schedule and picture boards, books on the subject of autism, internet web sites to explore) that we found to be successful, yet the materials are not being utilized. Many proven interventions are ignored.

Christine's vision for her "new life" in the group home was that she would be able to "come and go" like typical, normal 24 year-old girls. She is *supposed* to be assigned a one-on-one at all times, and with the use of our donated car, be able to go out into the community for the day if she chooses to. However, on weekends, much of her time is spent never leaving her group home. She has said that some of her weekend staff are "grouchy and they don't do fun things. They take me out shopping for an hour. That's it."\* If she needs assistance, she has to wait for a staff to be done with another client. We asked about her dedicated one-on-one on many occasions, but it is never addressed. The group home is not an "active" home. My assessment is that it is more along the lines of a nursing home.

This is a scenario that happens more times than not: all week long, Christine rarely goes out with her day program staff. On weekends, she looks forward to outings, but the outings are few and far between. Christine will then engage in a cycle of negative behaviors, which begets PRN's and probable restraints. All this is due to her lack of choices in her life.

So, many times, Jo-Ann and I try to support Christine on weekends because we realize she will become increasingly unhappy if she is not allowed to venture out and, in

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\* One weekend, her only outing was going to a 4:30 Sunday showing of the Jungle Book movie.



turn, her behaviors will deteriorate. (Additionally, the group home always seems to be short staffed on weekends.) I am sure staff will agree that when Christine is taken out into the community for an extended period of time, her behaviors are significantly improved when she returns to her group home.

The automobile we have donated to the group home has not been made available to my daughter or her group home for a year or so. My understanding is it is being used for another client. On Friday, April 22, 2016, I sent the below email to the Director of Business Operations, the Director of Residential Service and the group home manager regarding my concern:

To Whom It May Concern:

When my daughter, Christine [REDACTED] became a resident of the James L. Maher Jirmari Group Home, I donated an automobile to the James L. Maher Center for HER use. (Attached, is a copy of the letter I provided when the automobile title was signed over on July 24, 2014.) It is my understanding that the automobile has not been at the disposal for the staff at the Group Home to use nor has it been at the ready disposal for my daughters use. Further, I understand that on those rare occasions where the employees of the Jirmari home use it, staff is reprimanded for its use.

Per this letter, I would hope that the case of the donated 2004 Toyota Corolla is reviewed, discussed and turned over for the use of my daughter in the spirit of why it was donated I the first place.

Sincerely,  
Steven [REDACTED]

I received a reply from the Director of Business Operations where he wrote, "Your concerns will be passed on to the appropriate folks here at the Center. We will be in touch soon to give you a resolution." However, as of this writing, I have not been apprised of their decision.

A certain part of me wonders if the Maher agency agreed to take Christine as a client simply to have access to her high level of funding due to her Support Intensity Scale (SIS) evaluation. I was under the assumption that the Consent Decree directs our state to serve people with intellectual and developmental disabilities in the most integrated settings possible. Confining Christine to her home on weekends and not allowing her to spend at least 60% of her day program in the community is simply not in keeping with the spirit of the consent decree. If indeed the administration at the Maher Center is moving in the direction of community integration, I can assure you that it is at a snail's pace and at a detriment to my daughter's quality-of-life.

We were told that the preferred method of our communication with the James L. Maher Center should be through e-mail. Yet, most of our e-mails go unanswered. The manager of the day care program never returns calls or e-mails. On one occasion, I was told, in no uncertain terms, that Jo-Ann and I needed to "back off."

Of major concern is the seeming inability to properly staff group homes and day care providers. I am sure that this due to the relatively low pay which is needed to attract quality employees. My daughter is a very complex client. It takes staff who "get her;" staff who have a background and/or training in helping children and adults with autism. There are some overnights at the group home where there are only two staff members overseeing a house full of clients.

Another concern regards personal phone calls. Initially, we were in agreement that Christine's personal telephone calls to our home should be limited to one time a day that usually takes place at 7:00 PM. However, I question if this is a violation of her civil rights.

There is another major area of concern and it is that the system lacks a provision for checks and balances. Guardians are asked to sign quarterly purchase orders for services for their adult loved ones with special needs. The agency, in turn, bills for the services rendered. However, guardians/parents never see what is being billed for. On April 19, 2016, from 10:30 AM to 9:00 PM, Christine was with Jo-Ann and I. Also, on Sunday April 24, 2016, Jo-Ann was with Christine attending the Autism Walk at Goddard Park. There were no staff members (i.e.: one-on-one) accompanying her on either occasion. As a parent, though, I will never know if a bill was submitted requesting re-imbursement for services for these time periods.

I have found my daughter's state social worker, and her supervisor, are caring and attentive individuals. They have always been there when we needed them, in both the good times and not-so-good times. I would like to think that we have a very good working professional relationship. Sadly, though, it seems that there are limitations as to what type of interventions they can provide.

In spite of it all, though I must state emphatically that Christine absolutely loves her group home. She loves where it is located and she loves certain staff members. She wants to live there "forever," which is what she was told by an administrator at the James L. Maher center back in 2013 when she first visited the Jimari house.

In the area concerning her day program, my wife, Jo-Ann, has spent a lot of time and effort reaching out to other agencies. We have identified a day program that is very interested in having Christine as a client and we have since scheduled a meeting between all parties involved in our hope to "make it happen." Naturally, it will take a coordinated and cooperative effort among Christine's group home, her current day program placement and the new agency. Cooperation is the key word here because, without it, the new day placement will not work.

In closing, I write you today because I want my concerns and frustrations documented. Much of what I have written in this letter has already been shared in e-mails to various administrative people at the James L. Maher Center. More often than not, though, my efforts seem to be futile. I understand you do not represent an individual; you represent a department. However, the system in our tiny state of Rhode Island is broken.

My reason for writing you is not to be a thorn in anyone's side. Rather, it is to offer a perspective on what my wife and I have encountered in our years spent caring for our daughter with severe special needs. I hope you will listen and you will support all parents of adults with developmental disabilities. My wish is that we can work together to "find a fix" for the system that has a responsibility for caring for our beloved adults with special needs. They are our most vulnerable population and need our help, understanding, intervention and support.

After 21 years of caring for my daughter, I can assure you that I have no hidden agenda. I do not stand to gain financially in advocating for my daughter. I believe most parents of adults with special needs have honorable intentions. We want our loved ones to be treated as people with *abilities* and given the right to make life-style choices. We want to provide the voice for those who do not have the ability to advocate for themselves. Parents cannot do it alone and nor should the state. It truly needs to be a partnership, with perhaps the scale tipping slightly to the side of the parents, as no one knows our adult children better than we do.

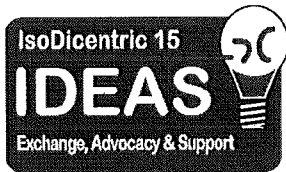
Sincerely,

Steven [REDACTED]  
[REDACTED]

(401) [REDACTED]

CC: United States Department of Justice  
Civil Rights Division  
Attn: **Nicole Kovite Zeitler**  
Trial Attorney,  
Disability Rights Section  
950 Pennsylvania Avenue, NW  
6027 NYA  
Washington, DC 20530  
202.616.4783  
[Nicole.Zeitler@usdoj.gov](mailto:Nicole.Zeitler@usdoj.gov)





## **Sudden Death in Chromosome 15q Duplication Syndrome** revised January 15, 2008 (changed text is underlined)

**Physician Advisory:** Please be advised that there is an increased risk of sudden, unexpected and currently unexplained death among children and young adults ages 7 and older with chromosome 15q duplication syndrome. The risk is approximately 1% per year. We are publishing this advisory so that physicians can be alert for potentially relevant symptoms and follow-up their patients according to their best clinical judgment with this information.

**Description of Sudden Death Cluster:** In the 21 month period between April 2006 and December 2007, IDEAS learned of the sudden, unexpected, and as yet unexplained deaths of six seemingly healthy young people with chromosome 15q duplication syndrome. In this same time period, another three children with duplications of chromosome 15q11-13 who were medically fragile because of their involved neurological status passed away.

The six cases of sudden and unexpected death involved young people of both genders between the ages of 7 – 26. All died during the night while they were in bed, presumably asleep. Parents reported hearing nothing alarming during the night. These young people were described by their parents, therapists, and doctors as lively, energetic, and affected by the cognitive disability, autism, and ADHD that are common with chromosome 15q duplication syndrome. Five of the six young people had recognized seizure disorders. One had no recent seizures, and three had seizures that were described as well controlled at the time of death, and one had not had a seizure for over a month.

**Potential Causes under Consideration:** There are three potential causes for these sudden deaths under consideration. The first consideration is whether any of these young people could have experienced a sudden respiratory event such as aspiration or hypoventilation. These events may or may not have been related to a seizure that was unwitnessed. Obstructive sleep apnea is another possibility, given the hypotonia that is common with duplications of chromosome 15q11-13.

The second line of inquiry is the possibility of a cardiac event. There are no documented congenital cardiac problems in chromosome 15q duplication syndrome. However abnormal heart rhythm, myocardial infarction, or cardiomyopathy cannot be ruled out.

The final possible cause under consideration is mitochondrial dysfunction. In 2003, Pauline Filipek, MD, published a case finding of mitochondrial dysfunction in autistic patients with 15q duplications (Annals of Neurology, 53(6), 801-804).

### **Recommendations:**

#### Treatment:

Although we do not have sufficient evidence to specifically connect any medication treatment to contribution to any of the deaths, 4 of the 5 patients with sudden, unexpected, and unexplained death were taking medications that are agonists at the GABA-A receptor, either benzodiazepines, phenobarbital, or both together, or alcohol derivatives. Such medications are known to be sedating and can be of some risk for patients with respiratory problems, raising concerns about a respiratory cause of death.

This advisory should not be considered a recommendation for anyone to change their treatment. It is intended for physicians treating young people with chromosome 15q duplication syndrome. Patients with this syndrome often have difficulty to treat seizures and severe sleep problems requiring the use of medications that cause side effects and/or expose patients to adverse events. However, this is an alert that patients with chromosome 15q duplication syndrome may be at higher risk of sudden death due to respiratory complications of GABA-A receptor agonists, including: benzodiazepines, phenobarbital and related medications, and other medications that are ethanol derivatives. This is not a documented risk and the risk of changing treatment may be greater than continuation, so each physician will need to weigh risks and benefits carefully, as in all cases. Physicians and families are reminded that changes in these medications should be done under physician directives and keeping in mind that these medications should be gradually decreased if they are to be withdrawn to avoid withdrawal syndromes which may lead to serious complications.

**Assessment:** The IDEAS professional advisory board does not have specific additional assessments to recommend at this time, but doctors should look carefully at each child's medical history (especially any potential respiratory, cardiac or metabolic red flags and overall response to illness), physical examination, and relevant laboratory evaluations. The following additional investigations may be considered (among others):

**Cardiac echocardiogram** to rule out cardiomyopathy and left or right ventricular hypertrophy, infarct, or wall motion abnormalities, or other abnormal cardiac findings.

**Signal averaged electrocardiogram** is more sensitive than a routine ECG and is a helpful non-invasive screening tool for increased risk for sudden cardiac death.

**A 24 hour heart rhythm ("Holter") monitor** may help detect cardiac arrhythmias. It must be stressed that such abnormalities have not yet been identified in this population.

**An overnight Video-EEG recording** may be warranted in patients with seizures, since the occurrence of nocturnal tonic seizures accompanied by alterations of the respiratory rate are a well known described phenomenon in 15q dup syndrome (Neurology 1997, 48: 1081-1086). When possible, the addition of extra leads to monitor respirations, pulse oxymetry and EKG (as is done as part of a sleep study or polysomnography) would be ideal.

**Research Resources:** Research is underway to investigate sudden death in chromosome 15q duplication syndrome. Four of the families made a brain tissue donation at the time of their child's death. The Autism Tissue Program (ATP) is coordinating the neuropathological exams. There were two autopsies performed. The first autopsy report stated that no satisfactory anatomical explanation of the death was found. The second autopsy report has not yet been released. Comprehensive autopsies after a sudden death are critical in our search to determine what is happening to these young people.

Families are encouraged to consider whether they would be willing to request an autopsy and make a tissue donation at the time of their child's death, whether it is sudden and unexpected or at the end of a more normal life span. Autopsy reports and brain tissue are critical resources for ongoing research efforts. Advance planning for tissue donation and autopsy are strongly encouraged. It has been our experience that the families have to advocate strongly for comprehensive autopsies and need to understand that these are not routinely performed because of the known chromosomal abnormality that is present. Autopsies can be paid for by IDEAS. The Autism Tissue Program is the resource for families wishing to make tissue donations. Families can pre-register with the ATP on their website: <http://www.brainbank.org/>

IDEAS is collecting information from families who pursue additional cardiac, metabolic or sleep studies. Families are encouraged to contact Nicole Cleary, IDEAS Board Chair with their results

(normal or abnormal). Physicians can assist in our research efforts by forwarding de-identified patient information to Nicole. It will be forwarded to the IDEAS professional advisory board.

Nicole Cleary, M.S.S.  
IDEAS Board Chair  
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**Final thoughts:** Please refer to the IDEAS web site at [www.idic15.org](http://www.idic15.org) for more information about this syndrome and specifically about any additional information that becomes available about sudden unexplained death in this syndrome. IDEAS provides support to families raising children with duplications of chromosome 15q11-13. If you are working with a family who is concerned about sudden death or any other aspect of raising an affected child, you may want to direct them to IDEAS to gain additional support.

Information compiled by IDEAS Professional Advisory Board Members:

*Edwin H. Cook, M.D., University of Illinois at Chicago.*

*Carolyn Schanen, MD, PhD, Nemours Biomedical Research.*

*Agatino Battaglia, MD, DPed, DNeurol, Stella Maris Clinical Research Institute for Child and Adolescent Neuropsychiatry, Pisa, Italy.*

With additional input from IDEAS Members:

*Vicki Miller, Cardiovascular and Thoracic Surgery, Brown University Program in Medicine*

*Nicole Cleary, M.S.S., IDEAS Board Chair*

## Quarterly Purchase Order for Individuals Receiving 24 Hour Residential Supports

## Under the Agency Model

This document is intended to be used by individuals and their families to select the supports they want to purchase in support of their Individualized Service Plan. This is not a service package. A Purchase Order must be submitted to BHDDH for approval before Service Package Tier can be assigned.

Name of Participant Submitting this Purchase Order Christine [REDACTED]  
 Social Security Number [REDACTED]  
 Current Address [REDACTED] Newport, RI 02840  
 Anniversary Date 1-Jul Residence Type Residential Supports  
 Purchase Order for the period: From Date 10/01/15 To Date 09/30/16

## PART A: Support Coordination is required in each individual's Personal Budget

	Enter Units	Unit Increment	Approved L9 in \$
The agency you have selected to work with is.....	<u>Maier Center</u>		
Support Coordination Modifier/Units of Service (Col F)	<u>Tier E (TG)</u>	<u>3</u>	<u>Month</u>
The name of your selected Support Coordinator is ...	<u>Josette Sobers</u>		

## PART B: Residential Supports

	Enter Units	Unit Increment	Approved L9 in \$
The agency you have selected to work with is.....	<u>Maier Center</u>		
24 Hour Residential Modifier/Units of Service (Col F)	<u>Comm. Res. Tier E</u>	<u>90</u>	<u>Days</u>
The street address where you will reside is .....	<u>228 Carroll Ave Newport, RI 02840</u>		
(For SLA only) The host family member's name is ...			
(For SLA only) Professional Supports		<u>15 min</u>	
SLA Respite		<u>15 min</u>	
Emergency Respite		<u>Days</u>	

## PART C: Day Activities

Make your selection(s) related to Day Activities.

Enter only on those lines where you are making a service request. Leave the remaining lines blank.

	Agency Selected to Provide Service	Tier/Ratio Selected by Individual	Enter Units	Unit Increment	Approved L9 in \$
Transportation to/from Day Activity	<u>Maier Center</u>	<u>Tier D, E (6-7)</u>	<u>128</u>	<u>Trip</u>	
Transportation to/from Day Activity				<u>Trip</u>	
Center-Based Day Program	<u>Maier Center</u>	<u>Tier E (1:1)</u>	<u>614</u>	<u>15 min</u>	<u>\$ 3,250.00</u>
Center-Based Day Program				<u>15 min</u>	
Community-Based Day Program	<u>Maier Center</u>	<u>Tier E (1:1)</u>	<u>922</u>	<u>15 min</u>	
Community-Based Day Program				<u>15 min</u>	
Home-Based Day Program				<u>Day</u>	
Professional Services While at Day Program	<u>Maier Center</u>	<u>1:1</u>	<u>65</u>	<u>15 min</u>	
Professional Services While at Day Program				<u>15 min</u>	
Prevocational Training				<u>Hour</u>	
Prevocational Training				<u>Hour</u>	
Job Coaching				<u>15 min</u>	
Job Coaching				<u>15 min</u>	
Job Development or Assessment				<u>Hour</u>	
Job Retention				<u>Month</u>	

By placing an X in box to the right, I/we are requesting quarterly authorizations equally throughout the year. Otherwise purchase orders for each time period must be submitted with the IISP/Annual Plan.

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Signature of Participant (or Legally Authorized Representative)

Date (month/day/year)

Signature of Representative from Agency #1

Date (month/day/year)

Signature of Representative from Agency #2

Date (month/day/year)

Signature of Representative from Agency #3

Date (month/day/year)